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HISTORICAL NOTE

Some contributions of Duchenne de Boulogne (1806–75)

PROGRESSIVE MUSCULAR ATROPHY

In 1849 Duchenne described a patient with spreading progressive muscular atrophy that started in the hands and spread slowly to the arms and legs, with no sensory signs, pain, or sphincter disturbance. Characteristically self effacing, he did not publish the case himself, but passed on his observations to François Amilcar Aran, physician to the Hôpital Saint Antoine. Aran published the paper[1] and acknowledged: "I owe a thousand thanks to my friend Duchenne de Boulogne who freely put at my disposal all his material . . ."

Duchenne's account (translated by G V Poore) followed in 1870[2]: "Muscles . . . often jerked by little fibrillary or partial contractions . . . agitated with worm-like movements. Progressive muscular atrophy attacks the upper limbs, and destroys its muscles in an irregular fashion. It begins in such cases by attacking one after another the muscles of the thenar eminence, spreading from the superficial to the deep layer. As soon as the abductor pollicis is wasted, its absence is marked by a depression, and by the attitude, during repose, of the first metacarpal bone, which lies too close to the second . . . Depressions of the hypothenar eminence and interosseal spaces next announce the atrophy of the muscles of those regions. The loss of the interossei muscles is shown by the claw-like attitude of the fingers . . . The atrophy may remain localised for many years . . . The flexors of the elbow and the deltoid are the first to atrophy. The triceps extensor cubiti is the last of the muscles of the upper limb to become affected. . . . Whenever all muscles of the arm have been atrophied, I have found a greater or lesser number of muscles of the trunk in the same condition . . . first, the lower half of the trapezius . . . I have usually seen the muscles of breathing and swallowing become affected. The atrophy equally invades the lower limbs, but only when the muscles of the upper limbs and trunk are in great part destroyed. It is most marked in the flexors of the ankle and hip. I have not seen atrophy attack both sides at once, but when one muscle is affected the corresponding muscles are usually attacked at no distant time."

He also wrote: " . . . (I have) only seen it begin in the lower limbs . . . twice out of 159 cases . . . in a good third of cases that electromuscular sensibility, as well as cutaneous sensibility was more or less weakened . . .. (There are) "change of form and attitude," superficial deformities, "functional troubles during voluntary action," and he notes the "wasting of intercostals and diaphragm . . . a great hindrance to breathing, and still more to phonation . . . there is no paralysis of the bladder or rectum . . . Duration.—This is very variable. The diseasemay reach its last stage in less than two years . . . I have seen it in this way remain localised for some eight or nine years . . . Electromuscular contractility is normal- . . . beyond doubt."

Pathologically, Duchenne reported the loss of striation, replacement by granular matter and fat vesicles, and fascicular atrophy of the muscles. But, he prefaces his account by saying that: "the feebleness of contractility . . . is chiefly the consequence of the wasting, . . . and not the result of paralysis, i.e. of a failure of the motor nerve action."

MUSCLE DISEASE AND "DUCHENNE'S DYSTROPHY"

His investigations of muscle disease continued with his invention of the "harpoon" that he employed to perform percutaneous muscle biopsies; not surprisingly, this aroused hostile criticism of its ethical propriety in the local press. The discovery of pseudo-hypertrophic paralysis, or myo-sclerotic paralysis in 1868,[3] was however, a remarkable and important contribution, dependent on and illustrated by pictures of histology obtained by harpoon biopsy: "This disease is mainly characterised: 1. By feebleness of movement, usually situated at first in the muscles of the lower extremities and of the lumbar spine, ultimately spreading progressively to the upper limbs, and increasing in intensity till all movement is lost; 2. increase in size of most of the paretic muscles; 3. By increase of the interstitial connective tissue of the paretic muscles, and in the more advanced stages by an abundant production of fibrous tissue or of fatty globules. The name I have given to this disease pseudohypertrophic muscular paralysis . . . has reference to the symptoms . . .. It may be called myo-sclerotic paralysis, a name which is more scientific and justified by pathological anatomy."

Of his many other contributions were original descriptions of the use of photography of microscopic histology, tabetic locomotor ataxia which contemporaries had confused with Friedreich's disease, the anterior horn cell lesions, which caused acute poliomyelitis, and glossolabio-laryngeal paralysis (bulbar palsy).

Guillaume Benjamin Amand Duchenne[4] was the son of a long lineage of seafarers and fishermen in the region of Boulogne sur Mer. According to Laségue and Strauss, he was of the middle height, thickset, active in movement, slow of speech and retaining to the last a faint provincial accent. He studied medicine in the University of Paris, under Laennec, Cruveilhier, and Dupuytren. He graduated in 1831. He returned to Boulogne to a limited private practice, but was badly affected by his young wife's death in childbirth. He lived only for his patients and for his scholarship.

Lonely and isolated from his friends, he returned to Paris in 1842 and started to experiment with Faradic current on the function of skeletal muscle. He sought no formal appointment, but attended patients in many Parisian hospitals, questioning and examining patients with laborious obsession, often following their progress by visiting them in their homes for many years. At times, he was humiliated by established physicians: "the monarchs of the wards", he called them. His reputation slowly increased, despite a neglect of pathological anatomy, and his dependence on his own observations rather than on neurological writings. Later in life he concentrated more on the nervous system than on muscles, taking up histology with youthful zest. Both Charcot and Trousseau fostered his recognition and showed him great respect: Charcot's lectures contain frequent acknowledgement of his work.

Neither succinct as a writer, nor systematic in his work, his lengthy papers emerged slowly. His first, *L'Electrisation Localisée et de son application à la pathologie et à la thérapeutique*, was published in 1855, was well received, and encouraged more research and trials of electrotherapy; by 1872 it had achieved a third edition.[2] In 1862 his previously estranged son joined him in Paris. He started, at last, to gain international respect. This culminated in election to many medical societies throughout Europe. A final disaster occurred when his son died of typhoid fever in 1871, with grave and lasting effects on his personal life. He died of a cerebral haemorrhage in 1875.

His epitaph we can leave to Charcot who remarked : "How is it that one fine morning Duchenne discovered a disease that probably existed in the time of Hippocrates? Why do we realise things so late, so poorly, with such difficulty . . ..Because our minds have to take in something that upsets our original set of ideas . . ." A bas-relief in the Salpêtrière shows the doctor attending his patient, applying electrodes attached to a simple generator. The accompanying plaque reads:

> A. Duchenne (de Boulogne)
>
> Electrisation Localisée
>
> Physiologie des Mouvements
> Neuropathologie

Postscript: Edward Meryon (1807–) presented a paper to the Royal Medico-chirugical society on 9 December, 1851, which described two typical "Duchenne" families and one with Becker type dystrophy. He recognised them as primary diseases of muscle[5] and showed postmortem the typical "granular degeneration"[6]

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